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Solitary Extramedullary Plasmacytoma in the Retroperitoneum

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Solitary extramedullary plasmacytoma is an uncommon neoplasm and occurs most frequently in the upper respiratory tract. Herein, we reported a solitary extramedullary plasmacytoma in the retroperitoneum. A 28-year-old man presented with obstructive jaundice and a retroperitoneal tumor. Ultrasound-guided biopsy confirmed that the lesion was a plasma cell neoplasm. A detailed investigation showed that no other sites were involved. The tumor got a moderate reduction following local irradiation, and a complete remission was achieved after 12 courses of adjuvant chemotherapy. Therefore, the possibility of a solitary extramedullary plasmacytoma should be considered in the differential diagnosis of obstructive jaundice without a history of multiple myeloma. Am. J. Hematol. 58:235–238, 1998. © 1998 Wiley-Liss, Inc.

Key words: extramedullary plasmacytoma; retroperitoneum; obstructive jaundice; thrombosis

INTRODUCTION

Plasma cell neoplasms are classified into four groups: solitary plasmacytoma of bone, extramedullary plasmacytoma, multiple myeloma, and plasma cell leukemia [1]. Extramedullary plasmacytoma is relatively rare as compared with solitary plasmacytoma of bone and multiple myeloma [1–3]. Most cases of extramedullary plasmacytoma occur in the upper respiratory tract, oral cavity, or gastrointestinal tract [1,3,4]. In this report, we describe a solitary extramedullary plasmacytoma in the retroperitoneum in a young adult presenting with prominent obstructive jaundice and inferior vena cava thrombosis.

CASE REPORT

A 28-year-old male patient presented with abdominal pain, deep jaundice, tea-colored urine, and clay stool. Laboratory data were as follows: AST 96 U/L (normal range, 0–34), ALT 130 U/L (normal range 0–36), alkaline phosphatase 227 U/L (normal range, 28–94), total bilirubin 15.4 mg/dl (normal range, 0–1.3), direct bilirubin 8.7 mg/dl (upper limit 0.4), total protein 8.8 g/dl, albumin 3.6 g/dl, uric acid 5.1 mg/dl, calcium 8.7 mg/dl, lactate dehydrogenase 68 U/L, white blood cell count 8,000 cells/c.c. with normal differential count, and hemoglobin 12.5 gm/dl. Abdominal computed tomographic scan showed a large tumor of soft tissue density in the right retroperitoneum (Fig. 1A) with anterior displacement of pancreatic head and involvement of right kidney at the mediosuperior border. The extrahepatic biliary tree was occluded with dilatation of the intrahepatic biliary tree. There was encasement of inferior vena cava, right renal vein, and right renal artery by the tumor. Inferior vena cava thrombosis (Fig. 1B) was also seen. It measured approximately 10 × 12 × 20 cm in greatest diameter. Angiographic study revealed that the main feeding artery of the retroperitoneal tumor arose from the posterior pancreaticoduodenal artery (Fig. 1C). Ultrasound-guided biopsy showed sheet-like infiltrations of plasma cells of varying degrees of differentiation (Fig. 2). There were also several plasmablasts with a single prominent nucleolus. Mitotic figures were occasionally seen. The

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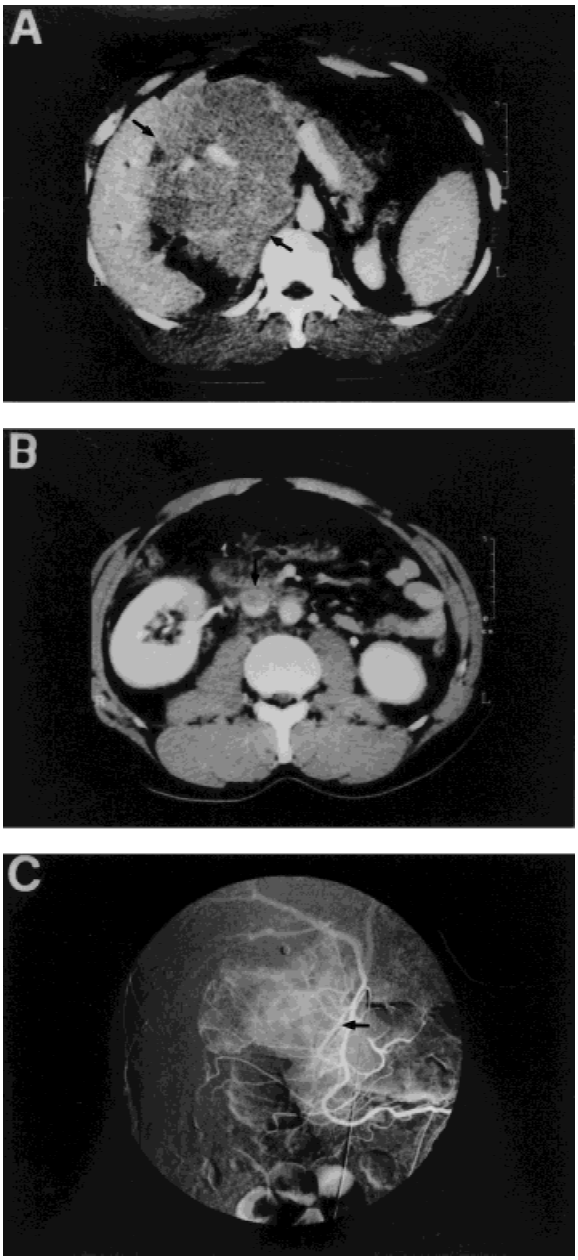

Fig. 1. Computed tomographic scan revealed a huge retroperitoneal tumor (arrows) of soft tissue density with anterior displacement of pancreatic head (A) and inferior vena cava thrombosis (arrow) (B). The main feeding artery arose from the posterior pancreaticoduodenal artery (arrow) (C).

plasma cells showed monotypic cytoplasmic immunostainings for IgG (1:500, Dako, Glostrup, Denmark) and kappa light chain (1:500, Dako), and were positive for VS38 (1:50, Dako), but negative for L26 (CD20) (1:50, Dako) and LCA (CD45) (1:100, Dako). Bone survey was negative. Bone marrow biopsy and aspiration revealed less than 2% plasma cells in bone marrow. Serum protein electrophoresis showed a monoclonal gammopathy with 3,100 mg/dl M protein. Immunoelectrophoresis showed a

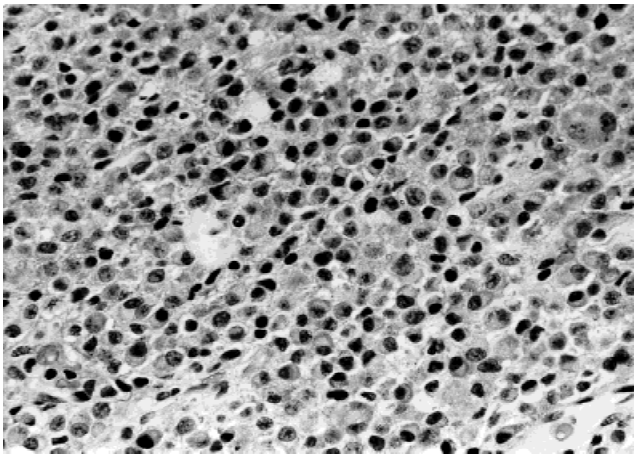

Fig. 2. Photomicrograph of the retroperitoneal tumor showed sheets of plasma cells of varying degrees of differentiation (hematoxylin and eosin, ×400).

kappa light chain spike in both blood and urine. The levels of immunoglobulins were as follows: IgG 4,685.0 mg/dl, IgM 135.7 mg/dl, IgA 172.3 mg/dl. Serum β_2-microglobulin was 3.474 mg/L. Therefore, the diagnosis of solitary extramedullary plasmacytoma of IgG-K type was made. During the period of work-up, total bilirubin rapidly rose to 25.6 mg/dl. The patient initially received radiotherapy with a total of 6,000 Gy in 29 fractions. A large residual mass measuring approximately 5 × 5 × 8 cm was still seen in the retroperitoneum (Fig. 3). Therefore, adjuvant chemotherapy with cyclophosphamide, vincristine, and prednisolone (COP) was given and achieved a complete remission after 12 courses of COP. Liver function tests returned to normal range, and protein electrophoresis and immunoelectrophoresis showed no paraprotein. To date, the patient has been free of disease for 1 year.

DISCUSSION

Extramedullary plasmacytoma is defined as a solitary tumor composed of monoclonal proliferation of cells with plasmacytic differentiation in an extramedullary site [5]. With solitary plasmacytoma of bone, they are viewed as a localized form of plasma cell neoplasm. They are associated with a much better prognosis than multiple myeloma [6]. Therefore, the diagnosis of localized plasmacytoma should be made only after detailed clinical, biochemical, hematological, and radiological investigations to exclude the possibility of localized presentation of multiple myeloma. Our case proved to be a solitary extramedullary plasmacytoma following a series of detailed examinations.

Solitary extramedullary plasmacytomas are relatively rare and account for less than 3–10% of all plasma cell neoplasms [7,8]. Most extramedullary plasmacytomas

Fig. 3. A large residual tumor (arrow) was still seen in the retroperitoneum.

were reported in the upper respiratory tract, including the oropharynx, nasopharnyx, nasal cavity, paranasal sinus, and larynx, followed by the gastrointestinal tract [1,3,4,8]. Extramedullary plasmacytoma occurring in the retroperitoneum was quite rare. Most reported cases were secondary to multiple myeloma [9]. In our investigation, we found only one case of solitary extramedullary plasmacytoma in the retroperitoneum reported in the English literature [10].

Obstructive jaundice was rarely diagnosed as a result of plasma cell neoplasms. Only a handful of cases were reported as due to extramedullary plasmacytoma of the pancreas [9,11–13] or duodenum [14] or the deposition of amyloid-like substances in the extrahepatic and large intrahepatic bile ducts [15]. However, all the cases occurred in patients with multiple myeloma. Thrombosis of the inferior vena cava was also an unusual presentation in plasma cell neoplasms. Only two cases with renal vein occlusion were reported [9,16]. One of them causes acute renal failure [16]. To our knowledge, our patient was the first case of solitary extramedullary plasmacytoma in the retroperitoneum presenting itself as obstructive jaundice and thrombosis of inferior vena cava.

The definite diagnosis of plasmacytoma was based on histological examination. The differential diagnoses included plasma cell granuloma, immunoblastic lymphoma, and lymphoplasmacytic lymphoma. The presence of many immature plasma cells and the demonstration of light chain restriction ruled out the possibility of plasma cell granuloma. The most important differential diagnosis in this case was non-Hodgkin's lymphoma. Some plasmacytomas of plasmablastic type may be misinterpreted as immunoblasticlymphoma since plasmablasts were very similar to immunoblasts [8]. Several cases of gastrointestinal plasmacytoma have been reinterpreted as low grade B-cell lymphoma with plasma cell differentiation [17]. It has been reported that failure to stain for CD45 and CD20 is highly characteristic for neoplastic plasma cells [18]. VS38 is a newly available monoclonal antibody for detecting plasma cell differentiation in routine sections [19]. The neoplastic cells in our case were positive for VS38 and negative for CD45 and CD20. The immunohistochemical study supported the diagnosis of plasmacytoma.

Extramedullary plasmacytomas are usually radiosensitive. Long-term local control can be achieved by adequate local radiotherapy [8]. In a review of the literature, approximately 94% of the cases achieved local control with doses in excess of 4,000 Gy [20]. Local failure with doses greater than 6,000 Gy was also reported in patients with extramedullary plasmacytoma [21]. There was still a large residual mass after completion of radiotherapy in this case. However, the size of residual mass was less than that of the original tumor and it presented a moderate reduction. In order to relieve a patient's symptom, we gave adjuvant chemotherapy to achieve a complete remission. The tumor may have been responding slowly to radiotherapy and complete response may have occurred with time and without chemotherapy. On the other hand, complete remission may be hastened by adjuvant chemotherapy. Therefore, the concurrent use of intensive, intermittent adjuvant chemotherapy with radiotherapy requires further study.

In summary, we reported the first case of extrahepatic biliary obstruction due to solitary extramedullary plasmacytoma in the retroperitoneum. Extramedullary plasmacytoma should be considered as the causes of obstructive jaundice even without a history of multiple myeloma.

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